HYDRAMNIOS AND CONGENITAL ANOMALIES

STUDY OF SERIES OF SEVENTY-FOUR PATIENTS

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A series of 74 patients with hydramnios was studied; 80% were multiparas and there were five sets of twins. There was a high rate of cesarean section (21.6%), breech deliveries (6.8%), diabetes (18.9%) and preeclampsia (14.9%). Among the 79 neonates delivered, there were 23 deaths and the perinatal mortality was 29.1%. There is a remarkable association of hydramnios and congenital anomalies; 21 infants (26.6%) had congenital anomalies and 14 (17.7%) had pathological conditions. Anencephaly and hydrocephaly were the most common lesions; 6 infants had gastrointestinal defects and 10 had anomalous lesions which would have obstructed swallowing or prevented the passage of fluid through the gastrointestinal tract while in utero. Special emphasis should be placed on lesions in which there is urgent need for correction of respiratory obstruction, early surgery, or both. Routine gastric catheterization and aspiration are therefore valuable diagnostic procedures.

Hydramnios has been associated with a high incidence of congenital anomalies and mortality in infants for many years (table 1). Recently this relationship has been suggested as a diagnostic sign in early recognition and guidance to the therapy of obstruction in the alimentary tract of the newborn infant.[1]

The purpose of this report is to present our experience with this disorder from 1953 to 1958 at the Sloane Hospital for Women and to emphasize a method of diagnosis which is used at this hospital to facilitate the discovery of gastrointestinal obstruction in all infants. In addition, the physiological processes involved in the normal and pathological formation of amniotic fluid are reviewed.

In data compilation, both mother and infant clinical records were used. Only those cases were accepted in which there was convincing clinical evidence of excessive accumulation of amniotic fluid. Two liters were taken as the upper limit of normal.[2] The cases of acute and chronic hydramnios were analyzed together.

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History

Incidence.—Collecting the amniotic fluid is difficult and uncertain and the resultant diagnosis is usually made solely on clinical impression. In milder cases of hydramnios the frequency of recognition will vary with the individual observer. For this reason the incidence of hydramnios varies greatly with different reports and ranges from 1 case in 62 deliveries [3] to 1 case in 754 deliveries [4] as shown in table 1. In 24,316 deliveries at the Sloane Hospital during the years 1953 to 1958 there were 74 cases of hydramnios, an incidence of 1 case in 329 deliveries. This frequency agrees closely with the statistics of the Johns Hopkins Hospital as reported by Eastman.[2] The combined figures of Ryder [5] in 1932, Yordan and D'Esopo [6] in 1955, and the present study total 78,616 deliveries at the Sloane Hospital for Women, with 391 cases of hydramnios, a frequency of 1 case in 201 deliveries.

TABLE 1.—*Perinatal Mortality and Congenital Anomalies Associated with Hydramnios*

Author	Cases, No.	Incidence*	Infants, No.	Perinatal Mortality, %†	Congenital Anomalies, %‡
Poeck,[1a] 1923	103	..	136	60	12.5
Floris,[1c] 1923	224	..	256	29	9.0
Taussig,[7] 1927	32	1:200	35	44	20.0
Ryder,[5] 1932	113	1:177	129	50	4.0
Mueller,[4] 1948	66	1:754	70	56	29.0
Macaffee,[1a] 1950	147	1:82	172	56	46.0
Prindle,[8a] 1955	146	1:278	155	51	44.0
Yordan and D'Esopo,[6] 1955	204	1:168	219	44	36.0
Eastman,[2] 1956	178	1:358	200	48	15.5
Scott and Wilson,[1b] 1957	169	1:166	..	..	29.5‡
Lloyd and Clatworthy,[1a] 1958	76	1:232	76	..	16.0
Barry,[3] 1958	213	1:62	231	69	59.5
Present study	74	1:329	79	29.1	26.6
Average		1:373		48.7	26.7

* Based on total number of deliveries.
† Based on total number of infants.
‡ Congenital anomalies associated with interference with fetal alimentation.

Parity, Multiple Births, and Type of Delivery.—Eighty per cent of the 74 mothers were multiparas. A similar high incidence of multiparous patients was also reported by Taussig [7] in 1927. There were five sets of twins in the series, representing 6.8% of the cases. This is seven times the usual occurrence of twins in the general patient population of Sloane Hospital during this period. With the exception of Lloyd and Clatworthy,[1a] the high frequency of multiple pregnancies is apparent in all reported groups of cases.

The types of delivery were interesting because the rates of cesarean sections (21.6%) and breech deliveries (6.8%) were significantly higher than the usual incidences of 8.5% and 3.4% respectively. The high cesarean section rate reflects the large number of diabetic patients found in the series (table 2).

Associated Complications of Pregnancy.—Table 2 lists the associated complications of pregnancy found in the series. The 18.9% incidence of diabetes is striking when compared to the average frequency of 0.42% at this hospital. It will be noted that the incidence of preeclampsia is 14.9%. This is about three times the expected rate in our patient population. There were five patients with Rh incompatibility as evidenced by elevated agglutinin titers, erythroblastotic infants, or both. The frequent association of hydramnios with diabetes, toxemia and Rh incompatibility is well-recognized [8] but its relative significance to the pathological accumulation of amniotic fluid is speculative. Plentl [9] believes that the common denominator is a metabolic or mechanical placental pathological change which may embarrass the placental-fetal circulation.

Fetal Outcome.—Seventy-nine babies were delivered from the 74 patients in this series. There were 23 fetal deaths, including 16 stillbirths and 7 neonatal deaths; the over-all perinatal mortality was 29.1%. This percentage represents one of the lowest perinatal loss rates reported for this syndrome (table 1). Floris [10] in 1923 had an almost identical incidence, but he was dealing with a large number of mild cases. The reason for this low rate is not apparent from the analyzed data. Eleven infants of this group had congenital anomalies, 10 infants had pathological abnormalities, and 2 infants were premature.

TABLE 2.—*Associated Complications of Pregnancy in Seventy-four Patients**

Complications	Patients No.	Patients %	Total Incidence at Hospital %
Diabetes mellitus	14	18.9	0.42
Preeclampsia	11	14.9	4.5
Hypertensive vascular disease	2	2.7	1.6
Heart disease	2	2.7	1.5
Rh incompatibility	5	6.8	..
Psychiatric disease	4	5.4	..
Fibromyomas of uterus	2	2.7	..
Epilepsy	2	2.7	..
Anemia	2	2.7	..
Thyrotoxicosis	1	1.4	..
Late latent lues	1	1.4	..
Hypothyroidism	1	1.4	..
Intercapillary glomerulosclerosis	1	1.4	..
Kyphoscoliosis with atelectasis	1	1.4	..
Pyelonephritis	1	1.4	..
Hyperemesis gravidarum	1	1.4	..
Mucous colitis	1	1.4	..
Adrenal cortical hypofunction	1	1.4	..
Total	53	..	..

*In 48 of these patients, history, laboratory findings, and results were normal.

Forty-one of the infants (51.9%) were apparently normal, and all survived. There was a high total frequency (19%) of prematurity which is consistent with the reports of others.[6] This is two and one-half times the usual incidence at Sloane Hospital. Only 3 of the 15 premature infants had no other associated abnormality. One of these was a 250-Gm. nonviable fetus.

Congenital anomalies were present in 21 babies. These include anencephaly, hydrocephaly, microcephaly, spina bifida, mongol, volvulus, tracheoesophageal fistula with atresia esophagus, imperforate anus, cleft palate, congenital heart disease, pyloric stenosis, genitourinary disease, deformed extremities and agenesis of ears. Pathological conditions were found in 14 other infants. These include

maceration, erythroblastosis, tight nuchal cord, hyaline membrane disease, edematous (diabetes) and meconium aspiration. Multiple anomalies were found in eight infants. Anencephaly was the most common lesion, but its incidence did not approach Barry's[3] 38.1% or Prindle's[8a] 23.4%. Hydrocephaly was the next most frequent type of anomaly. There were six babies with gastrointestinal defects, including three cases of volvulus, of which one was due to an atresia of the upper jejunum. Interestingly enough, one case of pyloric stenosis was diagnosed and an operation was performed

TABLE 3.—*Pathological Abnormalities Associated with Hydramnios*

Type	Live Births	Stillbirths and Neonatal Deaths	Total
Maceration	..	6	6
Erythroblastosis	2	1	3
Tight nuchal cord	1	..	1
Hyaline membrane disease	..	2	2
Edematous (diabetes)	1	..	1
Meconium aspiration	..	1	1

when the infant was eight weeks of age. This may represent a coincidental finding. Altogether, an incidence of 13.5%, 10 infants had anomalous lesions which would either prevent swallowing or obstruct the passage of fluid through the gastrointestinal tract while in utero. Scott and Wilson,[16] on the other hand, found this to be true in 29.5% of their cases.

Physiology of Amniotic Fluid

Many theories to explain the formation of amniotic fluids have been proposed; however, only three have received serious consideration. A fourth theory would consider the fluid of mixed origin. The oldest and simplest explanation is to regard this fluid as a product of fetal urination. It is true that the fetal kidney excretes urine in utero, however, this probably represents only a very small portion of the total amount of fluid present.[11]

Another possible source of the fluid is the amniotic epithelium which has been shown to have secretory characteristics. Although the organic constituents of the amniotic fluid may be derived in this manner, the water itself must come from some more effective source in order to be produced in such large quantity.[9] The third possible source is transudation from the maternal blood, the amnion regulating the passage of fluid in a manner similar to that of the peritoneum and pleura which results in a dialysate or ultrafiltrate of plasma. However, recent experimental work cannot substantiate this theory.[9]

The amniotic fluid is not a stagnant pool. On the contrary, there is experimental evidence that the exchange of the water content is a rapid process. Vosburgh and others[12] injected deuterium oxide (heavy water) into a maternal vein, and by sampling the amniotic fluid for the appearance of the isotope, found that, on the average, 35.4% of the water of the amniotic fluid is replaced every hour by water from the maternal plasma. This finding indicates that the entire water content changes about every three (2 hrs. 54 min.) hours. Investigation by Plentl and his group,[9] with isotopic tracers, led to a quantitative formulation of the over-all exchange between mother, fetus, and amniotic fluid. They found a net circulation of 600 ml. per hr. from mother to fetus to amniotic fluid and back to mother. In this circulation the water and electrolytic constituents are exchanged at their own individual rates and not en bloc. The amniotic fluid, therefore, cannot be a transudate or a dialysate of maternal plasma. However, these experiments do not give specific information on the site of exchange but only describe its over-all mechanism of exchange.

The removal of the amniotic fluid is enveloped in as much speculation as its formation. There may be transudation from amniotic fluid to mother by way of the amnion, decidua, and placenta, or the fetus may swallow or inhale it.[9] Macaffee[13] believed that the only area capable of exchanging adequate amounts of fluid is the fetal lung. The evidence for this is the finding of lanugo hair in fetal alveoli at autopsy and the demonstration of fetal respiratory movements in utero. However, James,[14] in a recent review of this subject, concluded that the lungs may be a source of fluid rather than a site

TABLE 4.—*Congenital Anomalies Associated with Hydramnios*

Type	Live Births	Stillbirths and Neonatal Deaths	Total
Anencephaly	..	5	5*
Hydrocephaly	1	3	4
Microcephaly	..	1	1
Spina bifida	1	..	1
Mongol	1	..	1
Volvulus	..	..	3*
With atresia upper jejunum	1	..	..
With congenital bands of upper jejunum	..	1	..
With common mesentery and herniation of liver	..	1	..
Tracheoesophageal fistula with atresia esophagus	1	1	2*
Imperforate anus	..	1	1
Cleft palate	1	1	2
Congenital heart disease	2	1	3
Pyloric stenosis	1	..	1
Genitourinary disease	3	2	5
Deformed extremities	..	1	1
Agenesis of ears	..	1	1

*Congenital anomalies definitely associated with interference with fetal alimentation in utero.

of absorption and that respiratory movements, being stimulated by asphyxia, only occur abnormally in the later period of gestation.

The simplest removal of this fluid would be via absorption through the fetal alimentary tract and then return to the maternal circulation via the placenta. That the fetus swallows amniotic fluid is a well-established fact.[18] Plentl and Gray[9] on the basis of in vivo experiments and hydrodynamic models concluded that at least 25%, and probably more than 50%, of the water transfer from the amniotic fluid to mother is through the fetus as intermedium. To what extent this is accomplished by fetal deglutition is unknown.

Etiology of Hydramnios

The most obvious explanation for hydramnios is that it is produced by either an overproduction or a deficient removal of amniotic fluid. The transfer of water is of such magnitude that a small difference of only a few milliliters per hour in the exchange rate could result in the clinical syndrome of hydramnios.

The close association of hydramnios with congenital anomalies (table 1), particularly those anomalies in which there is some interference with fetal deglutition or alimentation,[15] indicates a causal relationship. This leads to the hypothesis that a failure of the fetus to remove amniotic fluid, due to an obstruction of the passage of fluids in the proximal portion of the gastrointestinal tract (e. g., esophageal or duodenal atresia) or inability to swallow (e. g., anencephaly), would result in an excessive accumulation of liquid. Anencephaly may also be associated with an increased transudation of fluid from the exposed meninges into the amniotic cavity.[7] On the other hand, this syndrome may not be found if there is only a partial anencephaly with a medulla present. This would allow swallowing to take place. The occurrence of an obstructive lesion which does not result in hydramnios may be due to its distal location permitting proximal absorption (e. g., ileal atresia, meconium ileus or imperforate anus). In addition, there may be an incomplete obstruction, a wide tracheoesophageal fistula (type C) or the rare type of esophageal atresia (type D) in which both segments of the esophagus communicate with the trachea, thereby providing a route by which the fluid can reach the lower gastrointestinal tract.[3] However, this simple and convenient theory does not account for the occurrence of this syndrome in the presence of congenital anomalies in which the passage of fluid is not blocked or in the large proportion of completely normal infants.

Plentl,[9] on the basis of his physicochemical investigations on water exchange, implicates the fetal circulation and placenta as being etiological factors. This could account for the frequent association of this entity with such maternal diseases as toxemia, diabetes, and Rh incompatibility, all of which are characterized by abnormal placentae.

With the dynamic exchange of fluid perhaps we are dealing with multiple sites for interference, any one or a combination of which could produce an imbalance of only a few milliliters per hour in the transfer rates, which would result in the syndrome of hydramnios.

Diagnosis of Delivery Room Anomalies

Regardless of the precise pathophysiological mechanism involved, the remarkable association of hydramnios with congenital anomalies cannot be denied. Twenty-seven per cent of all the infants reported on in the literature had anomalies (table 1). Special emphasis should be placed on those lesions in which there is urgent need for correction of respiratory obstruction, early surgical therapy, or both.

Since it is particularly true of the obstructive gastrointestinal lesions that the longer the condition goes unrecognized the worse the prognosis becomes, it would behoove us to study and make a definite diagnosis in these infants as soon as possible. Recently it has been recommended [1] that in order to avoid a delay in diagnosis a catheter should be passed into the stomach of the infant to demonstrate the continuity of the esophagus. In addition, a small amount of air can be injected, thereby providing a contrast medium for roentgenographic evidence of other obstructive lesions within three hours of birth.[14]

In 1949, Gellis, White, and Pfeffer [16] first directed attention to the large volume of gastric contents found in infants delivered by cesarean section of diabetic mothers. They proposed routine gastric suction in these infants as a prophylactic measure against future regurgitation and aspiration. Since 1952 it has been the policy at the Sloane Hospital for Women to catheterize and aspirate the stomach of every infant without regard to the route of delivery. This is performed as soon as the respirations of the infants are well established. A soft two-holed no. 12 Rusch rubber catheter is attached to a glass bulb (De Lee) with a one-way trap and a rubber suction mouth piece. If the tip of the catheter is not seen in the left half of the abdomen, a stethoscope is placed over the abdomen and a short puff of air is used to confirm its position. There is a threefold purpose for this routine catheterization. The first purpose, the possible prevention of regurgitation and aspiration, has been mentioned. However, perhaps the more important reasons are the ruling out of the presence of esophageal atresia and the measuring of the gastric contents. The average volume found in infants delivered by cesarean section is 7.2 cc. (range 0 to 50 cc.), in breech deliveries it is 4.3 cc. (range 0 to 15 cc.), and in vaginal vertex it is 5.7 cc. (range 1 to 20 cc.). If 25 to 50 cc. are aspirated, an obstructive lesion of the upper gastrointestinal tract should be suspected and the infant studied closely with flat plates of the abdomen, further emptying and measuring of stomach contents, meanwhile withholding fluids by mouth. However, if over 50 cc. are aspirated initially, x-rays are taken within the hour. The absence of gas below the pylorus indicates obstruction and necessitates early exploration.

During the time that this routine has been used, six infants born at Sloane Hospital were found to have intestinal obstruction during the neonatal period (table 5). Four of the six babies had 38 cc. or more fluid present on gastric aspiration while still in the delivery room. Of the two with small quantities of fluid, the one in case 6 may have had volvulus developing sometime after birth, since the child was well for the first 10 days of life. From these data it would appear that through the simple

technique of measuring the gastric contents immediately after delivery, 80% of the in utero intestinal obstructions may be diagnosed.

Routine catheterization has not been as successful in the early diagnosis of tracheo-esophageal fistula with esophageal atresia. In only two of five infants born with this condition in the six-year period was the condition discovered in the delivery room. This may represent errors in technique on the part of inexperienced personnel. This is well illustrated by a case of atresia that was missed by four separate people using as the only criterion of patency the depth to which the catheter entered the esophagus. The position of the catheter and the diagnosis were later confirmed by gently puffing down the tube.

Freeman and Scott [17] believe that leaving the stomach empty has a favorable effect in the prevention of regurgitation and aspiration, with the subsequent development of pulmonary complications. No such relationship has yet been found at this clinic.

TABLE 5.—*Results of Gastric Aspiration in Infants Born at Sloane Hospital, Between Late 1952 and 1958, with Intestinal Obstruction During the Neonatal Period*

Case No.	Pathology	Hydramnios	Fluid, by Gastric Aspiration, Cc.*	Operation	Outcome
1	Congenital bands of upper jejunum; with volvulus	present	50	Lysis of adhesions, enteroenterostomy	Died 13 hours after birth
2	Atresia of upper jejunum	present	50	Gastrojejunostomy 3 days after birth	Died 7 days post-operatively
3	Atresia of upper jejunum with volvulus	present	75	Operated on 3rd day after birth	Recovered
4	Partial atresia of 2nd part of duodenum	absent	38	Jejunoduodenostomy 2nd day after birth	Died 8 days post-operatively
5	Volvulus with common mesentery and herniation of liver and multiple congenital anomalies	present	3	Ileostomy 1 day after birth	Died 2 days post-operatively
6	Volvulus with malrotation of intestines	absent	†	Operated on twice for recurrent volvulus (21st and 29th day)	Died 25 days post-operatively

* Aspiration performed in delivery room.
† Small amount of meconium removed.

Summary

Seventy-four cases of hydramnios occurring at the Sloane Hospital for Women in the years from 1953 to 1958 were analyzed with reference to parity, twin births, type of delivery, complications of pregnancy, and fetal outcome. There was a significantly higher incidence of multiparas, twin pregnancies, cesarean sections, and breech deliveries in mothers with hydramnios than in the general patient population. The incidence of diabetes mellitus (19%) and preeclampsia (15%) was remarkably greater than the usual rates of 0.42% and 4.5% respectively. Forty-one of the 79 infants were apparently normal, and all survived. Congenital anomalies were present in 21 (26.6%) and pathological abnormalities in 14 (17.7%). The over-all perinatal mortality was 29%. A technique of routine gastric catheterization and aspiration for all infants was used as a method of diagnosis of congenital anomalies of the upper gastrointestinal tract.

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